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CASE REPORT

Eosinophilic granuloma: Presenting as a draining fistula

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Abstract A case report of an orbital eosinophilic granuloma presenting as a fistula in a 15-year-old boy is presented. The patient had a history of a painful right upper eyelid swelling which required drainage and was considered as an abscess. An increase in swelling occurred and a small fistula appeared in the area subsequently. Computed tomography scan revealed a large soft tissue lesion in right superolateral orbit having intracranial extradural extension with destruction of bony orbital margin. Fine needle aspiration biopsy of the lesion revealed eosinophilic granuloma. A complete excision of the lesion was performed by a brow incision and histopathological examination of the excised specimen confirmed diagnosis. Although upper eyelid area eosinophilic granuloma is known to occur, its presentation as a fistula is not known.

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1. Introduction

Eosinophilic granuloma is a benign and localized form of Langerhans's cells-histiocytosis (LCH). It may present as a solitary or multiple lesions primarily involving the skull and the facial bones. This disease predominantly affects the children and young adults. A solitary LCH may present as an eyelid mass with swelling, ptosis, or rapidly increasing proptosis. We report a case of orbital eosinophilic granuloma that initially presented as a discharging upper eyelid fistula which we believe to be, the first case report of its kind.

2. Case report

A 15-year-old male patient presented with a history of right-sided dull aching orbital pain of 4 months duration. Two months later the patient developed a gradually enlarging nodular swelling without inflammatory signs over the outer aspect of right upper eyelid with an associated progressive drooping of the right upper eyelid. There was no history of any associated fever, malaise, or trauma. An initial presumptive diagnosis of right upper lid abscess was made at a peripheral treating facility. The patient was put on intravenous antibiotics after drainage of the abscess. Subsequently, the patient developed increase in swelling with painless foul swelling discharge from the drainage site with an increase in right upper eyelid swelling and mechanical ptosis. On examination at our centre, pus emerging from a fistula present in the middle of inflamed right upper eyelid with no associated significant lymphadenopathy was noted (Fig. 1). The examination of globe after holding the eyelid, revealed 2 mm downward displacement and restriction of elevation.

Visual acuity was 20/30 in the right eye and 20/20 in the left eye. Slit lamp examination revealed a small leucomatous corneal opacity not in the visual axis in the right eye. Fundus examination did not reveal any other significant abnormality. Pupillary reactions were normal. Neurological examination was unremarkable.

Computed tomography scan of head and orbit revealed a large soft tissue lesion in right superolateral orbit with intracranial extra-dural extension and destruction of bony orbital margin (Fig. 2). Fine needle aspiration from the lesion revealed proliferative histiocytic cells having round to oval shape with uniform coffee bean nuclei dispersed with fair number of eosinophilic cells in the background along with some multinucleated giant cells suggestive of eosinophilic granuloma. The lesion was excised through a brow incision along with necrotic bone were identified and removed from the subdural and subperiosteal spaces and bone edges were curetted.

Histopathological examination of the excised biopsy specimen confirmed the findings of FNAC (Figs. 3 and 4). Immunohistochemical staining with S-100 was positive.

3. Discussion

LCH encompasses three related disorders: Eosinophilic granuloma, Hand-Schüller–Christian disease, and Letterer–Siwe disease. Orbital eosinophilic granuloma is a rare disease accounting for less than 1% of all orbital tumours (Feldman et al., 1985) Most of the descriptions of the LCH are based on small case series and case reports. Incidence of orbital involvement may be upto 23%, and half of them can present

Figure 1 External photograph of 15-year-old male who was presented with a discharging fistula.

Figure 2 Computed tomograph scan (coronal view) howing a soft tissue lesion in right superolateral orbit having intracranial extra-dural extension with destruction of bony orbital margin.

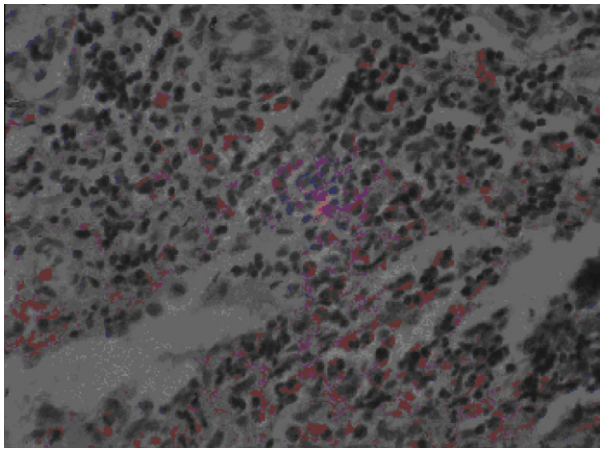

Figure 3 Histological section showing polymorphous population of proliferated histiocytes with coffee bean nuclei, plasma cells, lymphocytes and fair number of eosinophils (Haematoxylin & Eosin ×100).

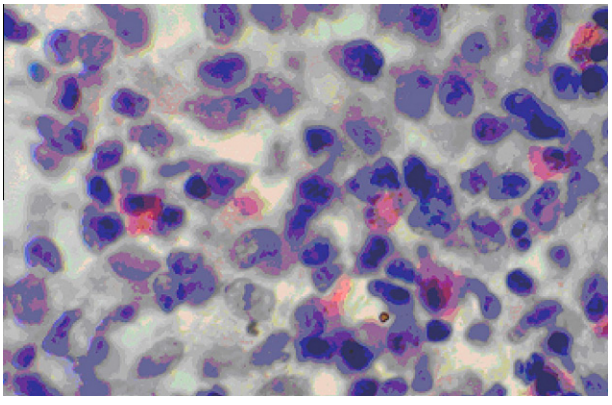

Figure 4 Higher magnification of the specimen showing classical morphology in the form of histocytes, plasma cells, lymphocytes and eosinophils (Haematoxylin & Eosin ×400).

with proptosis. The disease is characterized by unifocal or multifocal skeletal lesions and shows a male predominance with onset in the first or second decade of life. The common sites of involvement include frontal bone (Feldman et al., 1985), lateral wall of orbit (Filosom et al., 2008) and orbital roof. In rare cases, zygomatic sphenoid bone, and maxilla may be involved (Apple and Rabb, 1998). Eosinophilic granuloma may present as forehead and eyebrow swelling (Gerald and Kyung, 2003), painful eyelid swelling (Dubovy et al., 2003), ptosis, and rapidly increasing proptosis and in rare cases an abscess (Mehta and Shetty, 2001).

Bone pain and tenderness have also been reported. Radiologically this lesion is characterized by destructive lytic bone lesion, edges of which may be bevelled, scalloped or confluent (geographic), or show a ''button sequestrum''. It is assumed that the pathogenic langerhans cells liberate PGE2 and IL-1 to cause osteolysis.

Histopathological features of lesion include pathologic Langerhan's cells (histiocytes having densely pink cytoplasm with longitudinally grooved or cleaved nucleus), chronic inflammatory cells, giant cells, and eosinophils. Treatment strategies include conservative and surgical methods. Conservative approaches include intralesionalcorticosteroids, low-dose irradiation. Surgical methods include an incisional biopsy and curettage of the lesion.

Our patient had atypical clinical presentation of this rare disease. Although most of the clinical, pathological and radiological features in our case conform to the standard description of the disease, a discharging fistula following incision of suspected abscess to the best of our knowledge has not been reported to be the presenting feature of eosinophilic granuloma.

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